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NEURO-BEHÇET DISEASE WITH TYPICAL IMAGING FEATURES - A CASE REPORT

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ABSTRACT

Behçet's syndrome is named after Turkish dermatologist HulsiBehcet, who, in 1937, described a syndrome of recurrent aphthous ulcers, genital ulcerations, and uveitis leading to blindness. Radiologically, neuro-behçet disease (NBD) usually involves the white matter, brainstem, basal ganglia and thalamus. We report a case of Behcet syndrome with CNS manifestation with involvement of mesodiencephalic junction the most common neural parenchymal imaging feature described for the disease.

KEY WORD: Neuro- behçet disease, mesodiencephalic junction, parenchymal.

INTRODUCTION:

Behçet's syndrome is most common along the ancient Silk Road, extending from Asia to the Mediterranean. The central nervous system manifestations of the syndrome are termed as NeuroBehçet's disease (NBD), and has been described in 5% -50%,with the most common age group of presentation is 20-40 years(1,2,3). The exact pathophysiology of BD is not known but histopathologic findings has been characterized by nonspecific vasculitis involving various sized vessels in multiple organs.NBD show strong associations with human leukocyte antigen HLA-B51, genetic mutations including factor V Leiden(1,4). The BD has also been proposed to be a member of autoinflammatory family of disease(4).

The International Criteria for Behcet'sDisease (ICBD) was created in 2006 and now has replaced International Study Group Criteria (ISG criteria). In accordance with ICBD, the presence of any two of the following (genital aphthosis, skin lesions, eye lesions, and positive pathergy test) will diagnose/classify the patient as BD. For ICBD, vascular lesions were added, while oral aphthosis is no more mandatory. Getting 3 or more points diagnose/classify the patient as Behçet's syndrome (genital aphthosis 2 points, eye lesions 2 points, and the remaining each one point).Our patient has history of recurrent genital aphthosis, skin lesions with positive pathergy test hence fulfilling all the criteria of Behçet's syndrome (5).

The neurological involvement in Behçet's disease is either caused by primary neural parenchymal lesions (NeuroBehçet's syndrome, or brain stem meningoencephalitis) or non parenchymal involvement also known as neurovascular Behcet disease and it is usually associated with lesions secondary to dural sinus thrombosis(2,6,7,8).

Case report :

A 24 year-old male , follow up case of behcet disease with typical history of recurrent aphthous ulcers in the mouth and genital mucosa and positive pathergy test presented in medicine OPD with neurological signs and complained of numbness on the right side of the face, tongue and extremities and double vision for last 3-4 days .Rest of the systemic examination was normal. On basis of neurologic examination, patient was advised MRI brain.MRI brain showed T2 and FLAIR hyperintense signal involving predominantly left anterior midbrain in cerebral peduncle , left mesodiencephalic junction .signal changes were seen inferiorly extending to pons and superiorly till left side posterior limb of internal capsule(Fig. 1 A-1D and Fig. 2 A-2D).On coronal T2WI images lesions showing hyperintense signal seen continuous cephalocaudally as a long linear lesion, surrounded by edema in the presumed anatomical area of the corticospinal (pyramidal) tract(Fig.4A). Diffusion weighted and corresponding ADC images showed bright and low signal respectively suggestive of restricted diffusion (Fig.3A, 3B). No contrast enhancement was seen(fig.4A, 4B)

DISCUSSION:

The CNS manifestations of Behçet's syndrome can be categorized into Parenchymal and Non parenchymal group with parenchymal more common than non parenchymal. In one of case series the incidence of parenchymal form is 52% and that of non parenchymal form is 4%(8).The latter type is rarely complicated with the parenchymal lesions and should be called vascular-Behçet's disease and generally has a better prognosis compared with the parenchymal type(2,6,7). Rarely both vascular and parenchymal types can coexist(2,6,7, 8).

Parenchyma NBD usually affect the brainstem, mesodiencephalic junction, cerebral peduncle, hemispheral manifestations, spinal cord lesions, and encephalitic presentations. Non– parenchymal CNS involvement include dural sinus thrombosis, arterial occlusion, and arterial aneurysms. Among the parenchymal form the mesodiencephalic junction(MDJ) is the most common affected site and lesions of the MDJ junction in setting of appropriate clinical symptoms are highly suggestive of NBD (3,6,8,9,10).

MDJ lesions have further been characterized according to their location, the lesions which are located posteriorly at the MDJ tend to continue downward along the pontinetegmentum and superior cerebellar peduncle , whereas the lesions which are located anteriorly involve the corticospinal pyramidal tract. In our case also lesions were located anteriorly at MDJ and showed extension along presumed corticospinal pyramidal tract. The corticospinal pyramidal tract pathology on MR is manifested as asymmetrical longitudinal linear lesions extending from the posterior limb of the internal capsule down to the midbrain, pons and less frequently the medulla. Mid brain is more severely affected(6).

The lesions in NBD are described to show vasogenic edema and hence they usually dont show restricted diffusion on DWI, though its not uncommon to see restricted diffusion. Present case also showed bright signal on DWI and hpointense signal on corresponding ADC images consistent with restricted diffusion. The possible etiology of restricted diffusion has been attributed to acute inflammatory process due to vasculitis, with possibility of cytotoxic edema as cause of restricted diffusion has also been raised by few authors(11,12,13,14).

CONCLUSION:

To conclude this case report highlight the most common imaging features described for parenchymal NBD , showing characteristic involvement of MDJ region. Appropriate clinical and typical imaging findings help in diagnosis of NeuroBehcet disease.

Conflicts of interest: Nil

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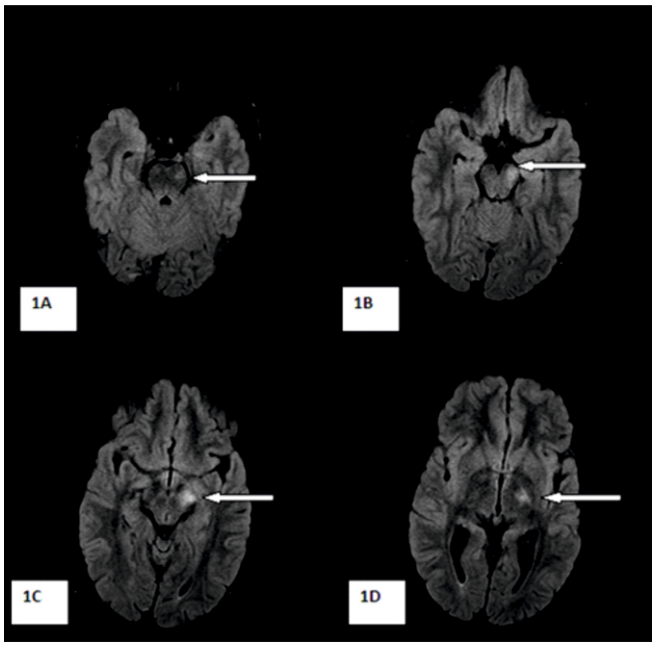
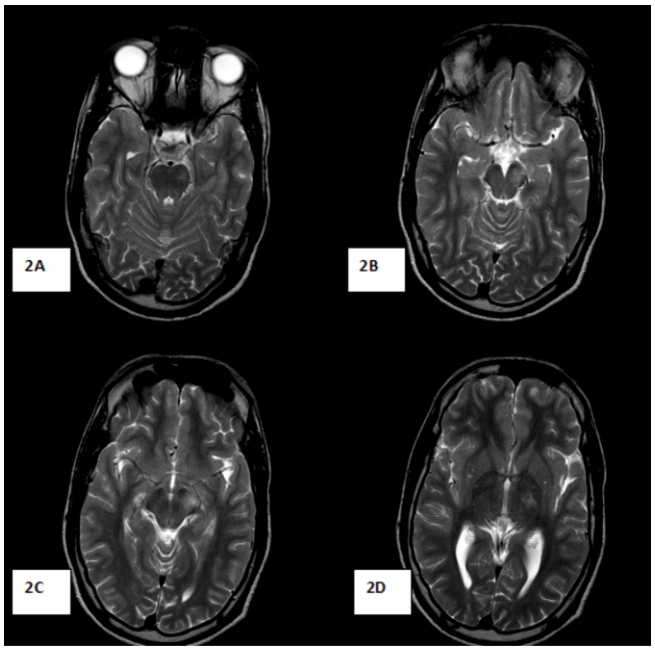


FIGURE 1: show FLAIR axial images with bright signal in pons **(FIG.1 A)**,anterior cerebral peduncle **(Fig.1B)**, posterior limb of internal capsule **(Fig. D)** with characteristic involvement of mesodiencephalic junction shown in **(Fig.1 C)**. Corresponding T2WI axial images with similar bright signal **(Fig. 2 ABCD)**.



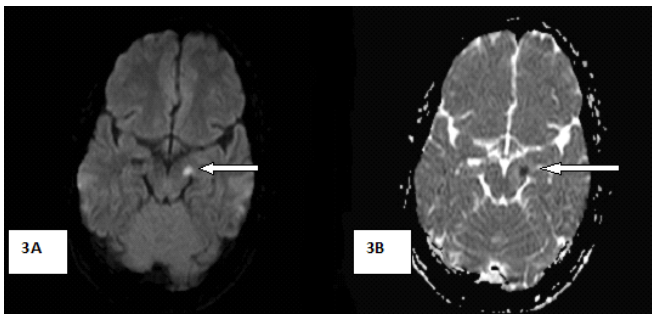


FIGURE.3A,3B. Axial weighted DWI and corresponding ADC images showing bright signal on DWI and low signal on ADC.

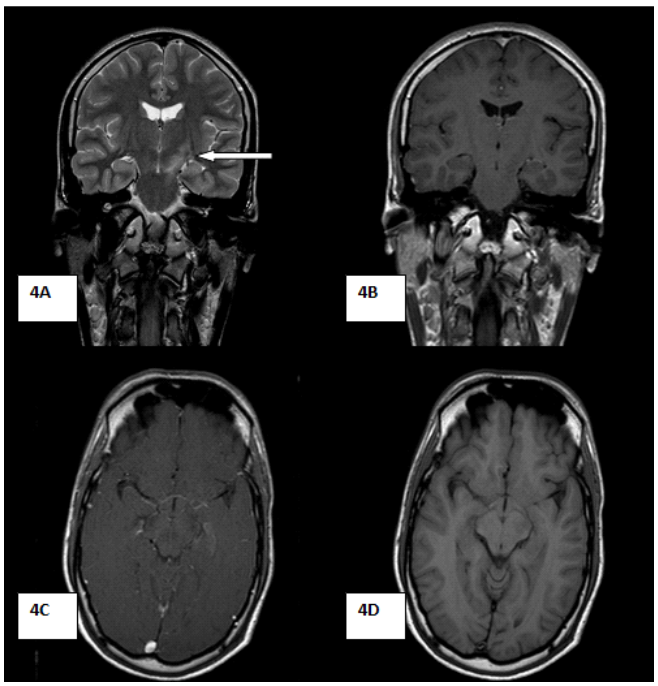


FIGURE.4. T2 Coronal image **(4A)** show that signal seen in axial images on T2WI and FLAIR are continuous cephalocaudally as a linear lesion, involving brainstem and MDJ region and located along the corticospinal (pyramidal) tract. **Fig.(4B & 4C)** are post I/V Gadolinium contrast enhanced images showing no enhancement, **Fig.4 D** show axial T1WI non contrast image for comparison.